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Cepacia syndrome in Czech CF patients

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Introduction: Cepacia syndrome (CS) is life-threating condition in CF patients colonized with bacteria belonging to Burkholderia (B.) cepacia complex. We experienced 8 cases of CS in last 10 years.
Case report: A 25-year woman with CF, homozygous for F508del, had chronic infection with Pseudomonas (P.) aeruginosa since 1984 and with B. cenocepacia since May 2003. In September 2004 she was admitted due to pulmonary exacerbation, without improvement after antibiotics (ATB). Because of high levels of total immunoglobuline (Ig) E (421.5 kIU/l) and positive titres of IgE anti-Aspergillus fumigatus, prednisone (40 mg daily) was administered. Subsequently, diabetes mellitus manifested during this therapy. In November 2004 CS developed and patient died within 39 hours after admission in spite of ATB and massive corticosteroid therapy. Serology for respiratory viruses and atypical bacteria was negative and decline of IgG levels and count of CD3+ lymphocytes (0.35 x 10^9/l) was found. Important parameters are summarized in a table:

Parameter	28 May 2003	20 Feb 2004	8 Oct 2004	14 Nov 2004
FEV_1 (% pred.)	55	46	32	n.a.**
BMI (kg/m^2)	19.2	19.2	18.0	15.4
CRP* (mg/l)	40.3	22.5	17.7	392.0
Albumin (g/l)	50.2	35.5	38.8	24.9
IgG (g/l)	25.3	20.8	20.3	14.3

*CRP…C-reactive protein, **n.a…not analysed

Conclusion: Changing properties of B. cenocepacia (influence of P. aeruginosa coinfection, oxidative stress or selectional pressure of ATB) and host „exhaustion" of immunity may contribute to CS pathogenesis. Presented case could give usefull guidance for future research on this terrible condition.
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Breath condensate pH levels in CF children with bacterial respiratory infection

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Introduction: Non-invasive monitoring of airway inflammation is important in CF children particularly in those with airway bacterial colonization. The pH values in exhaled breath condensate (EBC) has been proposed as a promising inflammatory and of oxidative stress. The aim of this study was to measure EBC pH as an evaluation of airway inflammation in CF children with or without bacterial infection, and for comparison, in healthy children.
Methods: We studied thirty CF children (mean age 9years) with P.aeruginosa or S.aureus respiratory infection, 10 CF children uncolonized and 10 healthy children.
Results: Data were expressed as means ±standard error of the mean. The pH values of EBC in the different groups were compared by analysis of variance with Mann-Whitney Rank Sum Test. A p value <0.05 was considered significant. Exhaled pH levels in all CF children groups were significantly lower in comparison to healthy subjects (7.76 ±0.26 vs 8.01 ±0.039; p=0.014).
No significant difference was however present among the EBC pH values of CF children (P.aeruginosa, S. aureus or non-colonized).
Conclusions: We conclude that pH values of EBC could be a useful indicator of oxidative stress in CF.

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Exhaled IL-8 and pH condensate levels in CF patients with respiratory exacerbation during antibiotic terapy

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Introduction: Interleukin (IL)-8 levels in exhaled breath condensate (EBC) is a promising non-invasive marker in acute exacerbation and the relationship between EBC pH, severity of disease and oxidative stress has also been studied.
Aim of our study as to assess whether exhaled IL-8 and pH levels in CF patients with respiratory exacerbation changed after antibiotic treatment.
Methods: fourteen CF patients (7 children, mean age 11years) with acute exacerbation were evaluated before and after antibiotic treatment.
Results: Data were expressed as means ± standard error of the mean. IL-8 levels and pH values before and after treatment were compared by paired t-test. A p <0.05 was considered significant.
Levels of IL-8 were decreased after antibiotic treatment, and pH condensate values were increased in all CF patients (tab.1).

Tab. 1: IL-8 levels and pH in EBC before and after antibiotic treatment.

EBC	Before treatment	After treatment
IL-8	0.36±0.028pg/ml	0.28±0.034pg/ml*
pH	7.61±0.084	7.36±0.087**

*(p=0.03); ** (p=0.04)

Conclusions: This study shows that exhaled IL-8 is a marker of acute pulmonary exacerbation and a reduced pH values can be considered as a marker of oxidative stress occurring in such situations.
These results suggest the application of EBC as a non-invasive monitor of the antibiotic treatment in CF patients.

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Exhaled condensate markers in cystic fibrosis children with bacterial airway infections

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Introduction: Leukotriene (LT) B4 and interleukin (IL)-8 are inflammatory mediators involved in the neutrophil response to bacterial infection. Aim of this study was to investigate whether the measurement of LTB4 and IL-8 in exhaled breath condensate (EBC) can reflect airway inflammation in CF children.
Methods: Thirty CF children (age mean 8years) were evaluated, 10 with P.aeruginosa, 10 with S.aureus, 10 uncolonized CF and 10 healthy subjects.
Results: Data were expressed as means ± standard error of the mean. The groups were compared by Scheffe test. A p<0.05 was considered significant. LTB_4 was higher in CF children with P.aeruginosa (49.8pg/ml+19.6) compared to those with S.aureus (2.7pg/ml+0.18;p=0.018), uncolonized (p=0.005) and healthy children (2.51pg/ml+0.18;p=0.005). IL-8 was elevated in CF children colonized by P.aeruginosa (0.72pg/ml+0.044) compared with all other CF subgroups (vs. S.aureus 0.56pg/ml+0.054;p=0.09, vs uncolonized 0.51pg/ml+0.052;p=0.017, vs normal subjects 0.36pg/ml +0.028;p=0.007). IL-8 levels were higher in CF children with S.aureus than in uncolonized CF (p=0.04) and in uncolonized CF compared with healthy subjects (p=0.027).
Conclusions: Our data support the application of the measurement of exhaled mediators in EBC as a non invasive method to evaluate airway inflammation and can represent a opportunity for a more targeted choice of therapeutic strategies in young CF patients.